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Research

Marchiafava-Bignami disease with rare Etiology: A Case Report

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ABSTRACT

Marchiafava-Bignami disease is a rare disorder of demyelination or necrosis of the corpus callosum and adjacent subcortical white matter that occurs predominantly in malnourished alcoholics[1]. The course of the disease may be acute, subacute, or chronic and is marked by dementia, spasticity, dysarthria, and inability to walk. Patients may lapse into coma and die, survive for many years in a demented condition, or occasionally recover[2]. Lesions appear as hypodense areas in portions of the corpus callosum on computed tomography (CT) and as discrete or confluent areas of decreased T1 signal and increased T2 signal on magnetic resonance imaging (MRI)[3]. Alcohol abusers without liver disease, amnesia, or cognitive dysfunction show thinning of the corpus callosum at autopsy[4] and on MRI[5,6], suggesting that alcohol or malnutrition damages the corpus callosum commonly in the absence of the necrotic lesions of Marchiafava-Bignami disease. These findings raise the possibility that aggressive nutritional supplementation along with a reduction in drinking can prevent the development of Marchiafava-Bignami disease in alcohol abusers. Here, we present a case report of MBD diagnosed in a 60 year-old male who presented with gait instability and slurred speech, in an effort to highlight the importance of obtaining MRI in patients presenting with behavioural disturbance and neurological findings, as well as discuss the relationship between thiamine supplementation and demyelinating diseases in the central nervous system.

Keywords: Marchiafava-Bignami disease, chronic alcoholism, Corpus callosum, Sandwich sign

INTRODUCTION

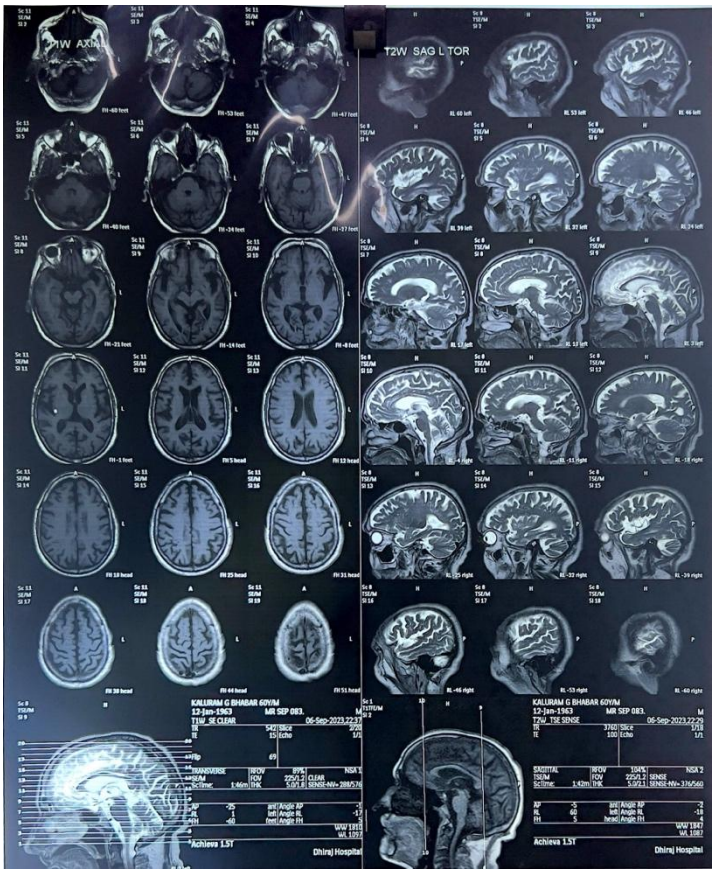
Marchiafava-Bignami disease (MBD) is a rare neurological disease often associated with chronic, heavy alcohol consumption and malnutrition, and is characterized by callosal lesions consisting of necrosis and demyelination (6–7). Over the past few years, magnetic resonance imaging (MRI) findings of the callosal and cortical lesions, which are critical for the diagno- sis of MBD, have been investigated (8). To date, the etiology of MBD is incompletely under- stood. In excess of 90% of patients with MBD exhibit a poor prognosis (9); however, with adequate therapy, these patients can recover completely, with the disappearance of the callos- al lesions on serial MRI (10–11).

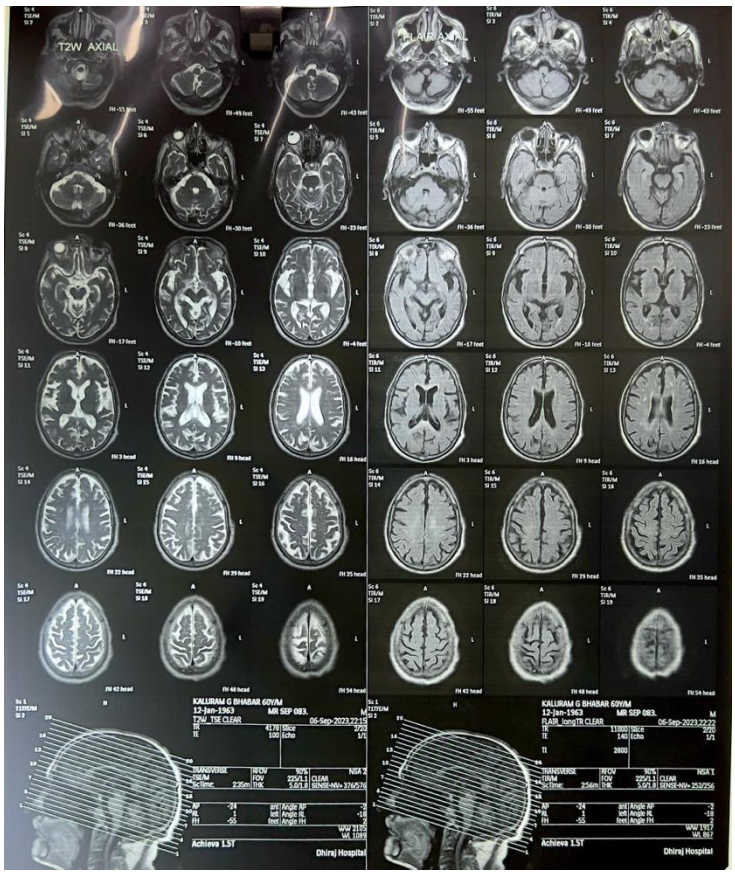
It is well known that MBD is widely observed and can be caused by any alcoholic beverage. It appears that, with alcoholism, the prognosis is worse; however, MBD can occur in patients who have no history of alcohol abuse (12–13). The main hypothesis for the pathogenesis of MBD is that the condition is associated with malnutrition or vitamin B deficiency (12), al- though there are reports describing cases of MBD caused by existing cyanide, CO poisoning and sepsis, as well as sickle cell disease and *Plasmodium* infection (14). The treatment of MBD in those patients remains controversial; however, the administration of nutritional fac- tors, such as high-dose group

B vitamins and folic acid, is believed to be beneficial, and nu- merous patients have shown improvements following the administration of vitamin B therapy (12,15).
The present study describes a case of MBD in a chronic alcohol drinker.

CASE PRESENTATION

A 60-Year old male patient came at our hospital with no known case of any co-morbidities, Presented with complaints of Bilateral lower limb weakness since 2 month followed by slurring of speech since 2 month and generalized weakness. The patient was relatively asymptomatic before 2 months after which he started to develop complains of bilateral lower limb weakness. It was gradual in onset and progression, associated with difficulty in walking, gait instability, difficulty in wearing slippers, unable to carry out daily routine activity, associated with easy fatigability. Patient also had the complain of slurring of speech since 2 month. It was associated with irrelevant speaking, cognitive impairment and poor calculation and memory and patient also had the complain of disturbance in sleep wake cycle. The patient had no significant past history of Fall or head trauma, chest pain, palpitation, perspiration, breathlessness, fever, headache, vomiting, seizure, Tuberculosis, Cerebral vascular accident, Diabetes Mellitus, Hypertension and also no history of any Drug abuse. Patients' personal history was significant as patient had reduced appetite, reduced sleep and Patient was Chronic Alcoholic and he use to take around 400 ml of country made alcohol daily and also smoker since 20-25 years and patient had last bout of alcohol 5 days back before being admitted to our hospital. On admission, the patient was drowsy but arousable, found to be malnourished and in altered sensorium (E3V4M5). His temperature was afebrile. Pulse was 110 beats per minute and regular , blood pressure was 100/70 mm of hg, Respiratory rate was 22 per minute. On general examination pallor, icterus, cyanosis, clubbing, lymphadenopathy, edema was not present . Signs of meningeal irritation including neck rigidity and kernig sign were absent. The bilateral pupils of the patient were round and equal, his light reflex and eyeball motion were normal, plantar were right sided flexor and left extensor, and The patient had no sensory disturbance, and his physiological reflexes were present without pathological reflex. The finger-to-nose and fast alternating movement tests showed the patient to be slightly clumsy. Laboratory results revealed normal hematological profile. Liver function test showed increased bilirubin. For Following presentation patient was advised for MRI (Plain & Contrast)





MR imaging in T2 and FLAIR Showed hyperintensity showing diffusion restriction on DWI noted along body, rostrum and genu of corpus callosum, Old lacunar infarcts noted involving bilateral frontal region. There are tiny T2 & FLAIR hyperintense areas in periventricular deep white matter region, possibility of periventricular chronic small vessel ischemic changes. There is mild prominence of sulci and gyri with mild dilatation of bilateral lateral ventricles, suggestive of cerebral cortical atrophy.

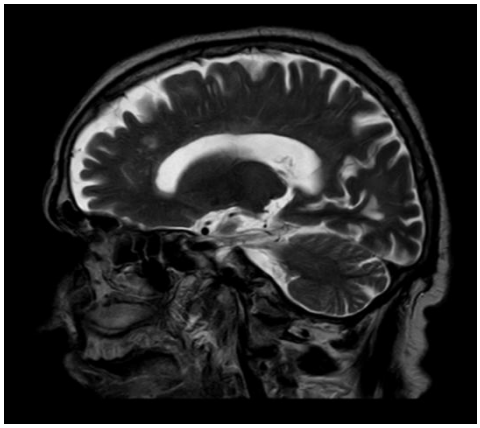


Fig : 1

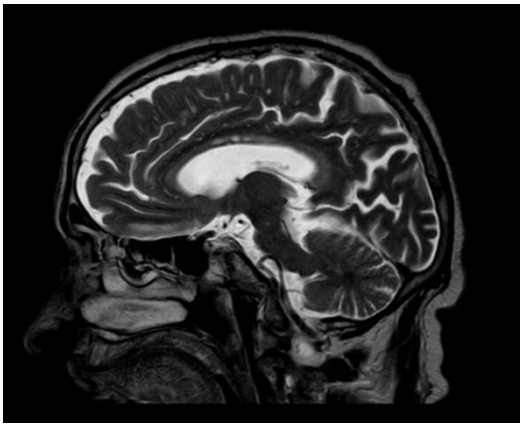


Fig : 2

Sagittal T2WI show low signal in the central layers of the genu, body & splenium of the corpus callosum with sparing of the dorsal and ventral layers producing the "sandwich sign" (fig :1,2)

And on the basis of this MR imaging finding various possible differential diagnosis can be made like, Cytotoxic edema, and Infarcts.

On the basis of history, clinical features and MR imaging findings the diagnosis of MBD was made. However, other lesions involving corpus callosum like multiple sclerosis, encephalitis, lymphoma, infarction, reversible lesion of corpus callosum were included in the differential diagnosis. History of chronic alchoholism with MR imaging findings favours the diagnosis of MBD in this case.

During the course of treatment, patient was told to consume a diet rich in vitamins to improve the brain blood and oxygen supply, and was prescribed Thiamine, Niacin, methylcobalamin, and folic acid and One weeks after admission, the slow responses and delusions of the patient had improved markedly, and patient was resolved from Bilateral lower limb weakness and was able to walk without instability in gait.

DISCUSSION

Marchiafava-Bignami disease (MBD) is a rare neurological complication of chronic alco- holism, with pathognomonic hallmark of corpus callosum demyelination [16]. It was first de- scribed by two Italian pathologists, Ettore Machiafava and Amico Bignami [17]. To date, the etiology of MBD is incompletely understood, as it is one of the rare complica- tions of chronic alcoholism (6–18). It is believed that MBD may be closely associated with chronic alcoholism and/or malnutrition caused by long-term alcohol abuse; however, MBD can also occur in patients who have no history of alcohol abuse (12–13). The main hypothesis for MBD pathogenesis is that the condition may be associated with malnutrition or vitamin B deficiency (12), although it is believed that MBD may have other etiologies. There are reports on MBD caused by existing cyanide, CO poisoning and sepsis, as well as sickle cell disease and Plasmodium infection (14).

Diagnosis is made on the basis of clinical findings in combination with radiological imaging features [17]. The corpus callosum appears hypoattenuated on CT, with the exception of cases that are characterized by subacute bleeding, in which it may be iso or hyperattenuated. The corpus callosum appears hyperintense on T2-WI, hypointense on T1-weighted images and proton density-weighted MR images during the acute phase. During the subacute phase, cystic lesions and small foci of T2 hypointensity can develop, most likely because of hemo- siderin deposition. In chronic stage signal intensity alterations become less evident but resid- ual atrophy of the involved structure is seen [19].

Marchiafava-Bignami disease may present in various clinical forms. Acute Marchiafava-Big- nami disease may present with seizures, impairment of consciousness, and rapid death. Suba- cute Marchiafava-Bignami disease

present with variable degrees of mental confusion, dysarthria, behavioral abnormalities, memory deficits and impairment of gait. Chronic Marchiafava-Bignami disease, is characterized by mild dementia which is progressive over years [20].

The recent clinical and neuroradiological classification of MBD describes two subtypes. Type A: acute to subacute onset of consciousness impairment, pyramidal tract signs, limb hypertonia, seizures, hyperintense swelling of the corpus callosum on T2-weighted MR se- quences and is associated with poor prognosis. Type B: normal or slightly impaired level of consciousness, dysarthria, gait disturbance, signs of interhemispheric disconnection and hy- perintense lesions on T2-weighted MR sequences partially involving the corpus callosum. Type B has favourable prognosis and lesions may reverse suggesting an underlying oedema rather than demyelination [21].

Treatment of MBD is usually empirical and consists of multivitamins, corticosteroids, stabi- lization of plasma glucose, and supportive care. Early diagnosis and prompt appropriate management are critical in reversing the underlying pathophysiology in the early stage [16].

CONCLUSION

Marchiafava-Bignami disease is a complication of chronic alcoholism with various clinical manifestations which is often misdiagnosed and mistreated. But recent advance in neu- roimaging has made early diagnosis of MBD possible & helps in early initiation of treatment. Recognition of the imaging features of MBD is essential for radiologist for diagnosis.

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